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Case Report

Surgical repair of severe mitral valve regurgitation complicated by incomplete cor triatriatum

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ABSTRACT

A 69-year-old woman with exertional dyspnea was referred emergently to our hospital for further evaluation. Transthoracic echocardiography showed severe mitral valve regurgitation and moderate tricuspid regurgitation, which were thought to be the main cause of her heart failure. An electrocardiogram showed paroxysmal atrial fibrillation. Mitral and tricuspid repair and pulmonary vein isolation were scheduled. Intraoperative transesophageal echocardiography revealed a fibromuscular diaphragm and multiple ostia in the left atrium, strongly suggesting cor triatriatum. After left atriotomy, an incomplete transverse membrane was identified in the chamber. The membrane was resected and the mitral valve was repaired; then a tricuspid annuloplasty was performed, and the pulmonary veins were isolated bilaterally. Her postoperative course was uneventful. Cor triatriatum is a rare congenital anomaly, and in some cases is associated with mitral regurgitation. In patients with severe mitral regurgitation, we recommend preoperative transesophageal echocardiography to obtain a correct diagnosis. We should evaluate carefully moderate to severe mitral regurgitation without pulmonary hypertension or left atrial dilatation taking cor triatriatum into consideration.

<**Learning objective:** Cor triatriatum is a rare congenital anomaly and in some cases is associated with mitral regurgitation. In patients with severe mitral regurgitation, preoperative transesophageal echocardiography is recommended to get a correct diagnosis. We should evaluate carefully moderate to severe mitral regurgitation without pulmonary hypertension or left atrial dilatation taking cor triatriatum into consideration.>

Introduction

Cor triatriatum is a rare congenital cardiac anomaly in which a fibromuscular membrane divides the left atrium into two distinct chambers. [1,2]. The hemodynamics of cor triatriatum is similar to that of mitral valve stenosis as a result of inflow obstruction by the intra-atrial membrane [3]. Here, we describe a patient with severe mitral valve regurgitation complicated by incomplete cor triatriatum, which was incidentally detected by intraoperative transesophageal echocardiography (TEE).

Case report

A 69-year-old woman was referred emergently to our hospital for exertional dyspnea and admitted for further evaluation of severe congestive heart failure. Three years before admission, she had a stroke. Eleven months before admission, mitral regurgitation and paroxysmal atrial fibrillation were diagnosed at an outpatient clinic. She was being treated with diuretics and antiplatelet drugs.

A chest X-ray showed significant cardiomegaly: the cardiothoracic ratio was 72%. An electrocardiogram showed paroxysmal atrial fibrillation, no ST-T changes, and left ventricular hypertrophy (SV1 + RV5 = 36 mV). Transthoracic echocardiography (TTE) revealed severe mitral regurgitation and moderate tricuspid regurgitation, which were thought to be the main cause of her heart failure. TTE also revealed a left ventricle of normal size, but the ejection fraction was only 37%, indicating decreased function. The left atrium was not dilated, the diameter was 41 mm. Since there was no evidence of torn chordae of the posterior mitral leaflet, the cause of mitral regurgitation was thought to be flail and prolapse of the lateral and mid portion of the posterior mitral leaflet. Pulmonary systolic pressure was 30 mmHg, diastolic pressure was 14 mmHg, mean pulmonary artery pressure was 21 mm Hg, and pulmonary capillary wedge pressure was 18 mmHg in cardiac catheterization.

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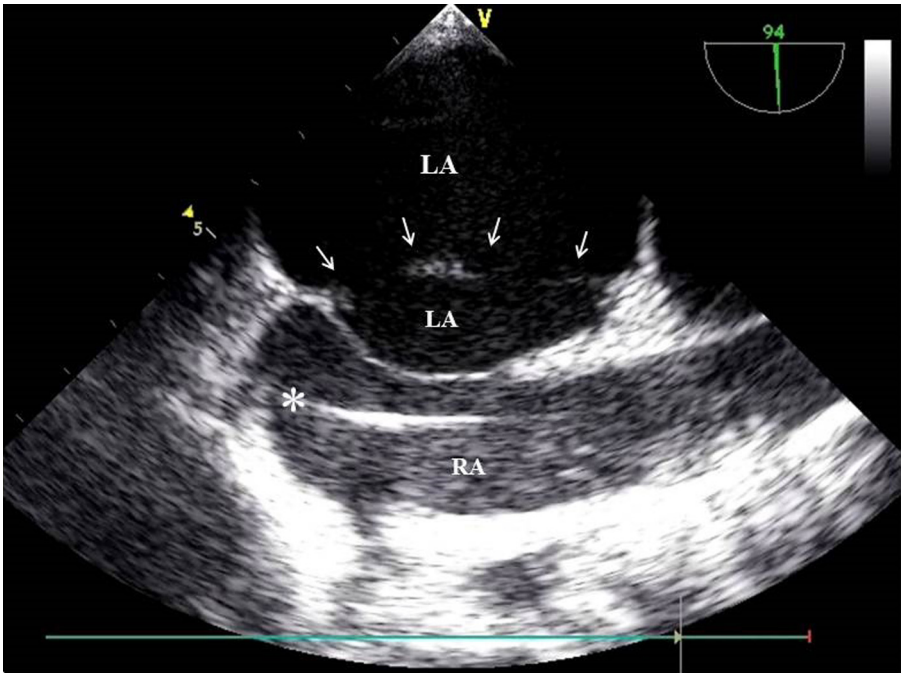

Fig. 1. Intraoperative transesophageal echocardiogram shows a fibromuscular diaphragm and multiple ostia in the left atrium. LA, left atrium; RA, right atrium; *, central venous catheter.

Mitral valve repair, tricuspid annuloplasty, and pulmonary vein isolation were scheduled.

After induction of general anesthesia, TEE revealed a fibromuscular diaphragm with multiple ostia in the left atrium (Fig. 1), strongly suggesting cor triatriatum. Surgery was performed through a median sternotomy: standard aortobicaval cannulation was used to establish cardiopulmonary bypass. After left atriotomy, an incomplete transverse membrane was identified and entirely resected. There was redundancy of the lateral and mid portions of the posterior mitral leaflet and small definite defect. The mitral regurgitation was controlled with mattress sutures and ring annuloplasty, a tricuspid annuloplasty and the pulmonary veins were isolated bilaterally. Intraoperative and postoperative echocardiograms revealed an entirely competent mitral valve with no regurgitation.

Her postoperative course was uncomplicated, and she was discharged on day 40 after a DDD pacemaker was implanted for bradycardia tachycardia syndrome. Postoperative TTE revealed improved cardiac function, the ejection fraction increased from 37% to 44% seven months after the operation. More than one and a half years postoperatively, she is followed at an out-patient clinic and free from recurrence.

Discussion

Cor triatriatum, one of the rarest congenital heart conditions, is identified in only 0.1% of patients with congenital heart disease [1–4]. In the classic form of cor triatriatum, the left atrium is separated into two compartments by a fibromuscular membrane. The clinical features are similar to those of mitral valve stenosis [1,3]. Pathologically, the obstruction caused by the membrane leads to a pressure gradient and increased pulmonary arterial and venous pressure. Cardiac catheterization typically reveals elevated pressure in the pulmonary arterial and capillary wedge positions that parallels the severity of the obstruction caused by the membrane. Although most cases of cor triatriatum are identified in infancy or early childhood, some are not detected until adulthood [1,3–8].

We present here an unusual case of mitral valve regurgitation complicated by cor triatriatum in an adult woman. The cor triatriatum was not identified by preoperative TTE. Rather it was an incidental finding during intraoperative TEE. Since our patient had neither pulmonary hypertension nor elevated pulmonary wedge pressure, we did not suspect other cardiac disease. In light of the unexpected finding of incomplete cor triatriatum, we investigated factors that warrant preoperative TEE to diagnose cor triatriatum in heart failure patients who have severe mitral regurgitation.

In patients with mitral regurgitation and cor triatriatum, the severity of hemodynamic abnormalities depends on the obstructive grade of the membranous diaphragm. Therefore, the obstruction is thought to be severe in early-onset cases, and mild in late-onset cases. Table 1 shows adult patients with mitral regurgitation and cor triatriatum reported in the literature [4–16]. Almost all reported cases were Lucas-Schmidt IA type of Cor triatriatum, which has no atrial septal defect, and three were IB type, which has atrial septal defect between the accessary chamber and the right atrium. We found that they were classified with age and mitral regurgitation grade. We divided the patients into three groups: those over 50 years of age whose mitral regurgitation was moderate to severe (Group 1), or mild to moderate (Group 2) and patients younger than 50 years of age (Group 3). There was no significant difference in gender distribution among the three groups. A large left atrium and atrial fibrillation were found only in Group 3, and a normal-size left atrium and normal sinus rhythm were found only in Group 1. Therefore, to get additional information in patients with severe mitral regurgitation, preoperative TEE might be recommended in younger patients with an enlarged left atrium and in the older patients with a normal-size left atrium and sinus rhythm.

The mechanism by which cor triatriatum exacerbates mitral regurgitation is unclear; however, it is thought that they are largely unrelated. Severe cor triatriatum occurs with mitral valve stenosis in infants; in reported adult cases, the main cause of mitral regurgitation is myxomatous change. In these patients, the left atrium may remain normal size because of disturbance of blood flow by the membranous diaphragm.

Table 1
Summary of reported literature about cor triatriatum complicated with mitral regurgitation.

Author	Year	Age	Sex	Type	Rhythm	EF	LAD	MR	PAP (m) (mmHg)	PCWP (mmHg)
Group 1										
Feld et al.	1992	55	F	IA	Af	50	Not enlarged	4	45/25/NR	28
Umemura	2013	69	F	IA	Paf	37	41 mm	4	30/14/21	18
Hogue et al.	1991	76	M	IA	Sinus	N/R	N/R	4	63/25/NR	N/R
Ludomiusky et al.	1990	65	M	IB	Sinus	N/R	Enlarged	3–4	60/40/NR	N/R
Tornic et al.	2011	52	F	IA	Sinus	55	N/R	3–4	N/R	N/R
Porter et al.	1983	52	F	IA	Af	N/R	Enlarged	3–4	65/33/45	N/R
Group 2										
Yamada et al.	2001	63	F	IA	N/R	N/R	53 mm	3	N/R	N/R
Okada et al.	1983	54	F	IA	Af	N/R	N/R	3	36/20/28	14
Leavitt et al.	1979	52	M	IA	Af	48	N/R	2	26/12/NR	14
Valakati et al.	2011	82	F	IA	N/R	N/R	N/R	N/R	N/R	N/R
Patel et al.	1990	74	M	IB	Af	N/R	N/R	N/R	60/24/NR	N/R
Kato et al.	2000	63	M	IA	Af	N/R	N/R	N/R	32/22/NR	19
Group 3										
Raggi et al.	1996	25	F	IA	Af	N/R	Enlarged	2	N/R	N/R
Fagan et al.	1991	37	M	IB	Sinus	N/R	Enlarged	N/R	N/R	N/R
Nagamori et al.	1989	33	M	IA	Af	N/R	N/R	N/R	48/35/42	35
Wong et al.	1989	33	M	IA	Af	N/R	Enlarged	4	N/R	18
Belcher et al.	1959	28	F	IA	Af	N/R	Enlarged	N/R	35/15/24	N/R
Lengyel et al.	1987	27	M	IA	Af	N/R	N/R	N/R	60/30/NR	N/R

Group1, patients over 50 years of age whose mitral regurgitation was moderate to severe; Group 2, those over 50 years whose mitral regurgitation was mild to moderate; Group 3, patients younger than 50 years.
EF, ejection fraction; LAD, left atrial dimension; MR, mitral regurgitation; PAP, pulmonary artery pressure; PCWP, pulmonary capillary wedge pressure; Af, atrial fibrillation; Paf, paroxysmal atrial fibrillation.
N/R, not recorded; MR grade: 4, severe; 3, moderate; 2, mild.
Lucas–Schmidt classification Type IA, which has no atrial septal defect (ASD); Type IB, which has ASD between the accessary chamber and the right atrium.

In conclusion, our patient with severe mitral valve regurgitation had an incomplete cor triatriatum, that was incidentally detected by intraoperative TEE. To obtain a correct diagnosis in patients with severe regurgitation, we recommend preoperative TEE. We should evaluate carefully moderate to severe mitral regurgitation without pulmonary hypertension or left atrial dilatation taking cor triatriatum into consideration.

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